

Anterior ethmoidal artery emerging anterior to bulla ethmoidalis: An abnormal anatomical variation in Waardenburg's syndrome

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ABSTRACT

In endoscopic sinus surgery, the anterior ethmoidal artery (AEA) is usually identified as it traverses obliquely across the fovea ethmoidalis, posterior to the bulla ethmoidalis and anterior to or within the ground lamella's attachment to the skull base. Injury to the AEA may result in hemorrhage, retraction of the AEA into the orbit, and a retrobulbar hematoma. The resulting increase in intraorbital pressure may threaten vision. Waardenburg's syndrome (WS) is a rare congenital, autosomal dominantly inherited disorder, distinguished by characteristic facial features, pigmentation abnormalities, and profound, congenital, sensorineural hearing loss. We present a case of AEAs located anterior to the bulla ethmoidalis in a 36-year-old male with WS and chronic rhinosinusitis. The anatomic abnormality was not obvious on a preoperative computed tomography scan and was discovered intraoperatively when the left AEA was injured, resulting in a retrobulbar hematoma. The hematoma was immediately identified and decompressed endoscopically without lasting complications. The AEA on the right was identified intraoperatively and preserved. The characteristic craniofacial features in WS were probably associated with the abnormal vascular anatomy. Endoscopic sinus surgeons should be aware of these potential anatomic anomalies in patients with abnormal craniofacial development.

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The anterior ethmoidal artery (AEA) is a branch of the ophthalmic artery. In endoscopic sinus surgery, it is usually identified in its location as it crosses the fovea ethmoidalis from posterolateral to anteriomedial, posterior to the bulla ethmoidalis, between the second and third lamellae (Fig. 1). The AEA may occasionally be located within the frontal recess in circumstances where the bulla ethmoidalis fails to reach the ethmoid roof and a suprabulla recess is present. Injury to the AEA may result in hemorrhage, retraction of the AEA into the orbit, and a retrobulbar hematoma. The resulting increase in intraorbital pressure stretches the optic nerve and reduces arterial bloodflow to the retina, threatening vision.

Waardenburg's syndrome (WS) is a rare congenital, autosomal dominantly inherited disorder, distinguished by characteristic facial features, pigmentation abnormalities, and profound, congenital, sensorineural hearing loss.¹

We present the first case of bilateral anterior ethmoid arteries emerging anterior to the bulla ethmoidalis in a patient with WS and suggest that these characteristic

craniofacial features may play a role in the abnormal vascular anatomy found.

Case

A 36-year-old male underwent bilateral endoscopic sinus surgery for recalcitrant chronic rhinosinusitis after failed medical therapy. The patient was known to have WS. Computed tomography (CT) sinus scans were examined before surgery, and it was noted that the AEA was difficult to identify (Fig. 2). During the procedure, at the time of the frontal recess clearance on the left hand side, the AEA was found to be anterior to the bulla ethmoidalis, within the frontal recess (Fig. 3), and was inadvertently injured. The injury was immediately identified as the left globe rapidly proptosed, and an endoscopic orbital decompression was undertaken. It was also noted intraoperatively that the right AEA emerged from the lamina papyracea anterior to the bulla ethmoidalis.

Postoperatively, the patient was reviewed by an ophthalmologist and admitted to the ophthalmology ward for overnight observation. He was discharged uneventfully the next day and was reviewed at 2, 6, 12, and 24 weeks postoperatively and made a complete recovery with no visual deficits.

DISCUSSION

Anterior Ethmoidal Artery

In endoscopic sinus surgery, the AEA may be identified on or just below the fovea ethmoidalis when an

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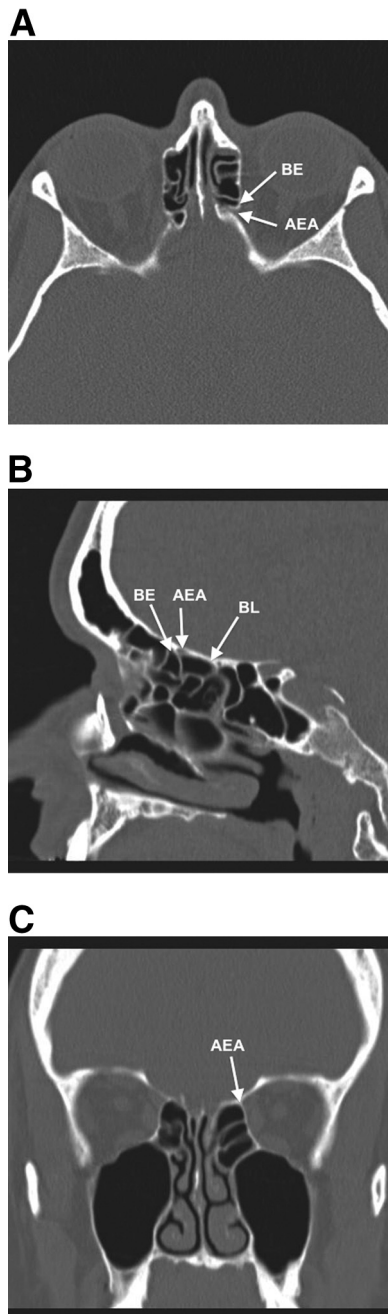


Figure 1. Axial, sagittal, and coronal views of left anterior ethmoid artery posterior to bulla ethmoidalis (normal). AEA = anterior ethmoidal artery; BE = bulla ethmoidalis; BL = basal lamella.

ethmoidectomy is being performed. Injury to the lateral part of the artery may result in retraction of the artery into the orbit, and if it continues to bleed, a retrobulbar hematoma may form.² A medial injury as the artery enters the lateral lamella of the cribriform plate may result in a cerebrospinal fluid leak. Accordingly, a detailed knowledge of the course and anatomic variations of the AEA is required to minimize the possibility of these complications occurring.

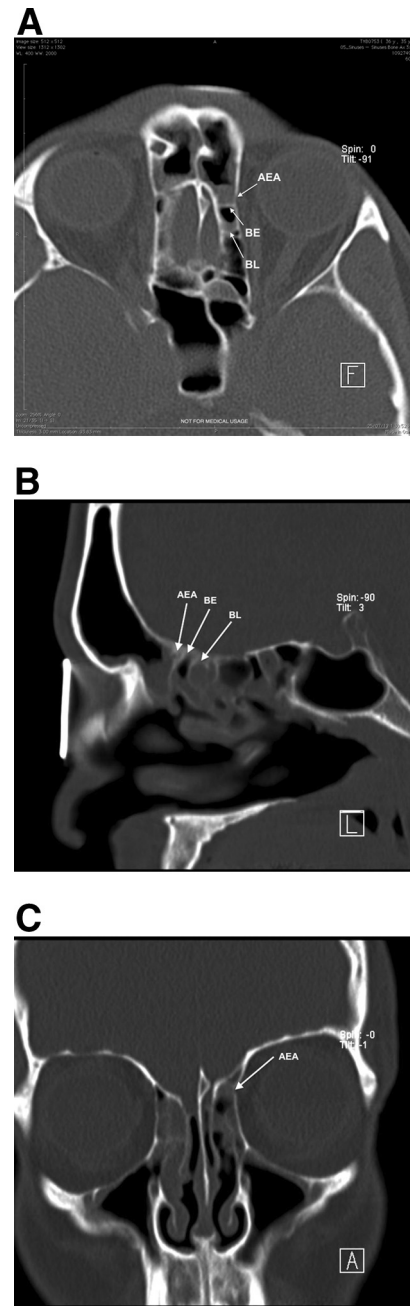


Figure 2. Axial, sagittal, and coronal views of left anterior ethmoid artery anterior to bulla ethmoidalis. AEA = anterior ethmoidal artery; BE = bulla ethmoidalis; BL = basal lamella.

The AEA originates from the ophthalmic artery in the orbit and passes between the superior oblique and medial rectus muscles. It enters the anterior ethmoidal foramen and then enters the anterior ethmoidal canal along with the anterior ethmoidal nerves. The artery then crosses the ethmoid sinus diagonally in an antero-medial direction to reach the cribriform plate, where it gives off an anterior meningeal branch. When the ethmoidal air cells are highly pneumatized, supraorbitally in particular, the process of pneumatization may enve-

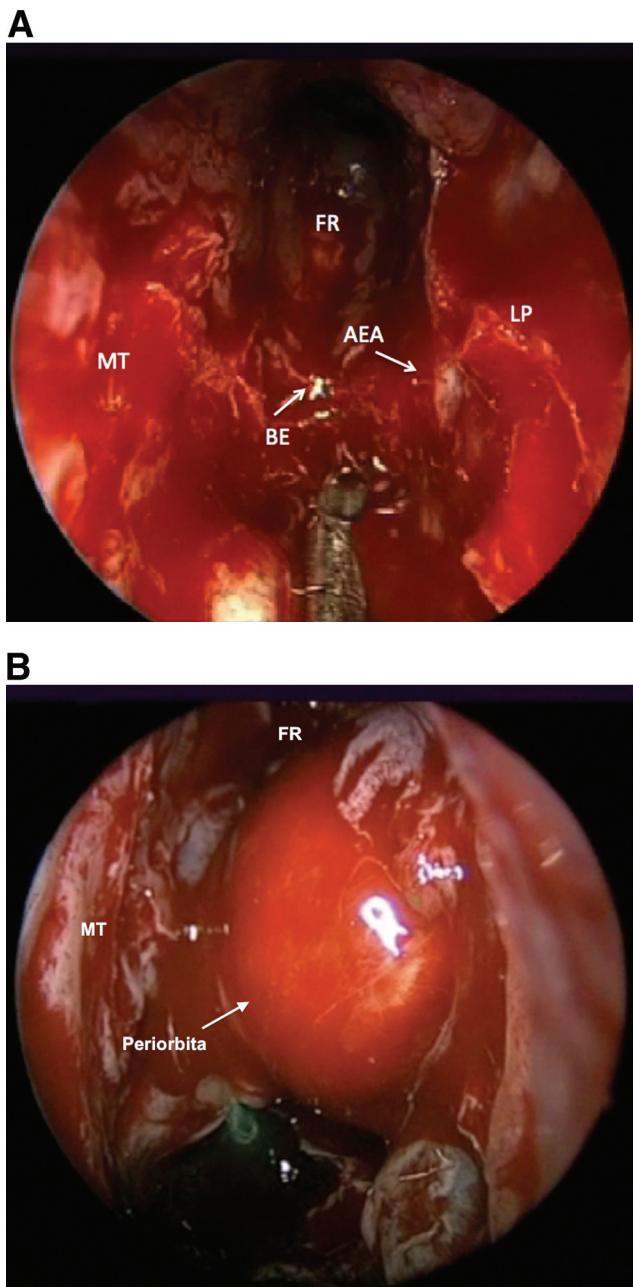


Figure 3. (A) Intraoperative view of arterial bleed from the left anterior ethmoid artery. (B) Orbital decompression. AEA = anterior ethmoidal artery; BE = bullae ethmoidalis; MT = middle turbinate; FR = frontal recess; LP = lamina papyracea.

loped the AEA so that its course is suspended off the fovea, usually in a bony mesentery or ground lamella. The AEA leaves the canal intracranially above the cribriform plate and enters the nasal slit on the lateral edge of the crista galli. The AEA then reenters the nasal cavity and divides into multiple branches, including the anterior septal branch and the anterior lateral nasal branch. Its terminal branch is the external nasal artery, which passes between the nasal bone and upper lateral cartilage to reach the nasal dorsum. The AEA supplies

the anterior ethmoid and frontal sinus, anterior third of the nasal septum, and the lateral wall of the adjacent nasal cavity.^{3,4}

Various landmarks have been used to enable endoscopic sinus surgeons to locate the AEA.⁵ These include: the junction of the posterior wall of the frontal recess and the adjacent anterior ethmoid cell where the artery is said to lie one to two millimeters posteriorly⁶; the axilla of the middle turbinate and the superomedial edge of the nose as these two points form a straight line to the AEA⁷; and the space in between the second (bullae ethmoidalis) and third (ground lamella) lamellae. In our case, we anticipated seeing the artery in between the bullae and the ground lamella. Failure to recognize its position anterior to the bullae resulted in the injury to the left AEA. This particular variation of the AEA has not been previously described in the literature.

Simmen *et al.* studied 34 cadavers and found that 85.3% of all AEAs were located in the recess between the second and third lamellae. The remainder were found in the retrobullar recess or roof of the bullae ethmoidalis.⁵ Moon *et al.*² also showed in their cadaveric study that 87.1% of AEAs were located between the second and third lamella. Eight (11.4%) AEAs were located within the third lamella, and one (1.4%) AEA was within the second lamella. In no cases did the AEA emerge anterior to the bullae ethmoidalis.² In Han *et al.*'s study, of 48 nasal cavities dissected, they found the AEA was located immediately anterior to (31%), at (36%), or immediately posterior to (33%) the ground lamella of the middle turbinate.⁸ Yang *et al.*⁴ evaluated 15 cadaver heads and documented AEAs found in the roof of the frontal recess (10.7%) and the roof of the posterior ethmoid sinus (3.6%). Eighty-five percent of the AEAs were posterior to the bullae ethmoidalis.

When the AEA is difficult to identify on CT scan, it may be helpful to repeat preoperatively the CT scan with a higher resolution.⁹ This would usually allow identification of an AEA, especially if it travels in a mesentery, and would reduce the possibility of iatrogenic injury. Intraoperatively, the AEA can then be identified and left intact. Alternatively, if an intact AEA prevents adequate dissection of diseased sinuses, once identified, preemptive ligation or cauterization with a bipolar diathermy could be undertaken. This process needs to be undertaken routinely in endoscopic procedures on the anterior skull base for which the fovea ethmoidalis needs to be removed. The technique for safe division of the AEA involves drilling of the bone of the floor of the canal to expose the artery and then diathermy of the artery with bipolar forceps. Both of these maneuvers are performed in the middle of the course of the artery across the fovea ethmoidalis to reduce the possibility of retraction into the orbit or injury to the lateral lamella of the cribriform plate.

In this case, retraction and continued bleeding of the AEA resulted in rapid proptosis of the eye. This problem was recognized immediately, and because the lamina papyracea was exposed, an endoscopic decompression of the medial orbital wall was able to be performed quickly. It is important to incise the periorbita to adequately decompress the orbit. If it is not possible to perform this procedure immediately, the orbit can be decompressed by performing a lateral canthotomy and cantholysis. An alternative route to decompressing the medial orbital wall is via a Lynch incision. The choice of surgical approach is governed by the situation and the available expertise. Retinal ischemia and visual loss may occur with as little as 60 minutes of raised orbital pressure, so a timely decompression is mandatory.¹⁰

Waardenburg Syndrome

Waardenburg's Syndrome is characterized by dystopia canthorum, a prominent broad nasal root, synophrys, a white forelock, and heterochromia iridis. The complexity understanding of WS continues to grow as the syndrome displays both molecular and clinical heterogeneity.¹¹ Currently, there are four subtypes of WS,¹² and six autosomal genes are implicated. A mutation of the Paired Box 3 gene is responsible for the majority,¹¹ resulting in a defect in neural crest migration and development.¹³ The association between the variation of AEA anatomy and WS has not been documented. Only one case report described a variant of WS with underdevelopment of the paranasal sinuses ranging from mild hypoplasia to aplasia.¹¹ Our patient had type 1 WS, which includes dystopia canthorum. His skull base was developmentally abnormal, including an unusual location of the AEA.

CONCLUSION

In this case, the emergence of the AEA anterior to the bulla ethmoidalis is a significant finding, because it reminds the endoscopic sinus surgeon of the potential for anatomic variability of the AEA. The characteristic craniofacial anomalies of WS probably played a role in

the abnormal vascular anatomy found in our patient. Endoscopic sinus surgeons should be aware of the potential for anatomic anomalies in patients with abnormal craniofacial development.

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